



General

Guideline Title

Spasticity in children and young people with non-progressive brain disorders: management of spasticity and co-existing motor disorders and their early musculoskeletal complications.

Bibliographic Source(s)

National Collaborating Centre for Women's and Children's Health. Spasticity in children and young people with non-progressive brain disorders. Management of spasticity and co-existing motor disorders and their early musculoskeletal complications. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Jul. 54 p. (NICE clinical guideline; no. 145).

Guideline Status

This is the current release of the guideline.

Regulatory Alert

FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- [December 14, 2016 – General anesthetic and sedation drugs](#) : The U.S. Food and Drug Administration (FDA) is warning that repeated or lengthy use of general anesthetic and sedation drugs during surgeries or procedures in children younger than 3 years or in pregnant women during their third trimester may affect the development of children's brains. Consistent with animal studies, recent human studies suggest that a single, relatively short exposure to general anesthetic and sedation drugs in infants or toddlers is unlikely to have negative effects on behavior or learning. However, further research is needed to fully characterize how early life anesthetic exposure affects children's brain development.
- [August 31, 2016 – Opioid pain and cough medicines combined with benzodiazepines](#) : A U.S. Food and Drug Administration (FDA) review has found that the growing combined use of opioid medicines with benzodiazepines or other drugs that depress the central nervous system (CNS) has resulted in serious side effects, including slowed or difficult breathing and deaths. FDA is adding Boxed Warnings to the drug labeling of prescription opioid pain and prescription opioid cough medicines and benzodiazepines.

Recommendations

Major Recommendations

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Collaborating Centre for Women's and Children's Health (NCC-WCH) on behalf of the National Institute for Health and Clinical Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

Principles of Care

Delivering Care

Children and young people with spasticity should have access to a network of care that uses agreed care pathways supported by effective communication and integrated team working.

The network of care should provide access to a team of healthcare professionals experienced in the care of children and young people with spasticity. The network team should provide local expertise in paediatrics, nursing, physiotherapy, and occupational therapy. Access to other expertise, including orthotics, orthopaedic surgery, and/or neurosurgery and paediatric neurology, may be provided locally or regionally.

If a child or young person receives treatment for spasticity from healthcare professionals outside the network team, this should be planned and undertaken in discussion with the network team to ensure integrated care and effective subsequent management.

Management Programmes

Following diagnosis, ensure that all children and young people with spasticity are referred without delay to an appropriate member of the network team.

Offer a management programme that is:

- Developed and implemented in partnership with the child or young person and their parents or carers
- Individualised
- Goal focused

When formulating a management programme take into account its possible impact on the individual child or young person and their family.

Carefully assess the impact of spasticity in children and young people with cognitive impairments:

- Be aware that the possible benefit of treatments may be more difficult to assess in a child or young person with limited communication.
- Ensure that the child or young person has access to all appropriate services.

Identify and agree with children and young people and their parents or carers assessments and goals that:

- Are age and developmentally appropriate
- Focus on the following domains of the [World Health Organization's International Classification of Functioning, Disability and Health \(children and youth version\)](#) :
 - Body function and structure
 - Activity and participation

Record the child or young person's individualised goals and share these goals with healthcare professionals in the network team and, where appropriate, other people involved in their care.

Help children and young people and their parents or carers to be partners in developing and implementing the management programme by offering:

- Relevant, and age and developmentally appropriate, information and educational materials
- Regular opportunities for discussion and
- Advice on their developmental potential and how different treatment options may affect this

Supporting the Child or Young Person and Their Parents or Carers

Offer contact details of patient organisations that can provide support, befriending, counselling, information, and advocacy.

Ensure that children and young people have timely access to equipment necessary for their management programme (for example, postural management equipment such as sleeping, sitting, or standing systems).

The network team should have a central role in transition to prepare young people and their parents or carers for the young person's transfer to adult services.

Monitoring

Monitor the child or young person's condition for:

- The response to treatments
- Worsening of spasticity
- Developing secondary consequences of spasticity, for example pain or contractures
- The need to change their individualised goals

The network of care should have a pathway for monitoring children and young people at increased risk of hip displacement.

Recognise the following clinical findings as possible indicators of hip displacement (hip migration greater than 30%):

- Pain arising from the hip
- Clinically important leg length difference
- Deterioration in hip abduction or range of hip movement
- Increasing hip muscle tone
- Deterioration in sitting or standing
- Increasing difficulty with perineal care or hygiene

Offer a hip X-ray to assess for hip displacement:

- If there are clinical concerns about possible hip displacement
- At 24 months in children with bilateral cerebral palsy

Consider repeating the hip X-ray annually in children or young people who are at Gross Motor Function Classification System (GMFCS) level III, IV, or V.

Consider repeating the hip X-ray after 6 months in children and young people where the initial hip migration is greater than 30%, and then consider repeating the hip X-ray every 6 months after this if the hip migration is increasing by more than 10 percentage points per year.

Physical Therapy (Physiotherapy and/or Occupational Therapy)

General Principles

All children and young people with spasticity referred to the network team should be promptly assessed by a physiotherapist and, where necessary, an occupational therapist.

Offer a physical therapy (physiotherapy and/or occupational therapy) programme tailored to the child or young person's individual needs and aimed at specific goals, such as:

- Enhancing skill development, function, and ability to participate in everyday activities
- Preventing consequences such as pain or contractures

Give children and young people and their parents or carers verbal and written (or appropriate formats) information about the physical therapy interventions needed to achieve the intended goals. This information should emphasise the balance between possible benefits and difficulties (for example, time commitment or discomfort), to enable them to participate in choosing a suitable physical therapy programme.

When formulating a physical therapy programme for children and young people take into account:

- The views of the child or young person and their parents or carers
- The likelihood of achieving the treatment goals
- Possible difficulties in implementing the programme
- Implications for the individual child or young person and their parents or carers, including the time and effort involved and potential individual barriers

When deciding who should deliver physical therapy, take into account:

- Whether the child or young person and their parents or carers are able to deliver the specific therapy
- What training the child or young person or their parents or carers might need

- The wishes of the child or young person and their parents or carers

Ensure that any equipment or techniques used in the physical therapy programme are safe and appropriate, in particular for children or young people with any of the following:

- Poorly controlled epilepsy
- Respiratory compromise
- Increased risk of pulmonary aspiration
- Increased risk of bone fracture due to osteoporosis (for example, those who are unable to walk, malnourished, or taking anti-epileptic therapy)

Encourage children and young people and their parents or carers to incorporate physical therapy into daily activities (for example, standing at the sink while brushing teeth in order to stretch leg muscles).

Specific Strategies

Consider including in the physical therapy programme 24-hour postural management strategies to:

- Prevent or delay the development of contractures or skeletal deformities in children and young people at risk of developing these
- Enable the child or young person to take part in activities appropriate to their stage of development

When using 24-hour postural management strategies consider on an individual basis low-load active stretching or low-load passive stretching.

Offer training to parents and carers involved in delivering postural management strategies.

Consider task-focused active-use therapy such as constraint-induced movement therapy (temporary restraint of an unaffected arm to encourage use of the other arm) followed by bimanual therapy (unrestrained use of both arms) to enhance manual skills.

When undertaking task-focused active-use therapy consider an intensive programme over a short time period (for example, 4–8 weeks).

Consider muscle-strengthening therapy where the assessment indicates that muscle weakness is contributing to loss of function or postural difficulties.

Direct muscle-strengthening therapy towards specific goals using progressive repetitive exercises performed against resistance.

Following treatment with botulinum toxin type A, continuous pump-administered intrathecal baclofen, orthopaedic surgery, or selective dorsal rhizotomy, provide an adapted physical therapy programme as an essential component of management.

Ensure that children and young people and their parents or carers understand that an adapted physical therapy programme will be an essential component of management following treatment with botulinum toxin type A, continuous pump-administered intrathecal baclofen, orthopaedic surgery, or selective dorsal rhizotomy.

Continuing Assessment

Reassess the physical therapy programme at regular intervals to ensure that:

- The goals are being achieved
- The programme remains appropriate to the child or young person's needs

Orthoses

General Principles

Consider orthoses for children and young people with spasticity based on their individual needs and aimed at specific goals, such as:

- Improving posture
- Improving upper limb function
- Improving walking efficiency
- Preventing or slowing development of contractures
- Preventing or slowing hip migration
- Relieving discomfort or pain
- Preventing or treating tissue injury, for example by relieving pressure points

When considering an orthosis, discuss with the child or young person and their parents or carers the balance of possible benefits against risks. For example, discuss its cosmetic appearance, the possibility of discomfort or pressure sores or of muscle wasting through lack of muscle use.

Assess whether an orthosis might:

- Cause difficulties with self-care or care by others
- Cause difficulties in relation to hygiene
- Be unacceptable to the child or young person because of its appearance

Ensure that orthoses are appropriately designed for the individual child or young person and are sized and fitted correctly. If necessary seek expert advice from an orthotist within the network team.

Be aware when considering a rigid orthosis that it may cause discomfort or pressure injuries in a child or young person with marked dyskinesia. They should be monitored closely to ensure that the orthosis is not causing such difficulties.

The network of care should have a pathway that aims to minimise delay in:

- Supplying an orthosis once measurements for fit have been performed and
- Repairing a damaged orthosis

Inform children and young people who are about to start using an orthosis, and their parents or carers:

- How to apply and wear it
- When to wear it and for how long:
 - An orthosis designed to maintain stretch to prevent contractures is more likely to be effective if worn for longer periods of time, for example at least 6 hours a day.
 - An orthosis designed to support a specific function should be worn only when needed.
- When and where to seek advice

Advise children and young people and their parents or carers that they may remove an orthosis if it is causing pain that is not relieved despite their repositioning the limb in the orthosis or adjusting the strapping.

Specific Uses

Consider the following orthoses for children and young people with upper limb spasticity:

- Elbow gaiters to maintain extension and improve function
- Rigid wrist orthoses to prevent contractures and limit wrist and hand flexion deformity
- Dynamic orthoses to improve hand function (for example, a non-rigid thumb abduction splint allowing some movement for a child or young person with a 'thumb in palm' deformity)

Consider ankle-foot orthoses for children and young people with serious functional limitations (GMFCS level IV or V) to improve foot position for sitting, transfers between sitting and standing, and assisted standing.

Be aware that in children and young people with secondary complications of spasticity, for example contractures and abnormal torsion, ankle-foot orthoses may not be beneficial.

For children and young people with equinus deformities that impair their gait consider:

- A solid ankle-foot orthosis if they have poor control of knee or hip extension
- A hinged ankle-foot orthosis if they have good control of knee or hip extension

Consider ground reaction force ankle-foot orthoses to assist with walking if the child or young person has a crouch gait and good passive range of movement at the hip and knee.

Consider body trunk orthoses for children and young people with co-existing scoliosis or kyphosis if this will help with sitting.

Consider the overnight use of orthoses to:

- Improve posture
- Prevent or delay hip migration

- Prevent or delay contractures

Consider the overnight use of orthoses for muscles that control two joints. Immobilising the two adjacent joints provides better stretch and night-time use avoids causing functional difficulties.

If an orthosis is used overnight, check that it:

- Is acceptable to the child or young person and does not cause injury
- Does not disturb sleep

Continuing Assessment

- The network team should review the use of orthoses at every contact with the child or young person. Ensure that the orthosis:
 - Is still acceptable to the child or young person and their parents or carers
 - Remains appropriate to treatment goals
 - Is being used as advised
 - Remains well fitting and in good repair
 - Is not causing adverse effects such as discomfort, pain, sleep disturbance, injury, or excessive muscle wasting

Oral Drugs

Consider oral diazepam in children and young people if spasticity is contributing to one or more of the following:

- Discomfort or pain
- Muscle spasms (for example, night-time muscle spasms)
- Functional disability

Diazepam is particularly useful if a rapid effect is desirable (for example, in a pain crisis).

Consider oral baclofen if spasticity is contributing to one or more of the following:

- Discomfort or pain
- Muscle spasms (for example, night-time muscle spasms)
- Functional disability

Baclofen is particularly useful if a sustained long-term effect is desired (for example, to relieve continuous discomfort or to improve motor function).

If oral diazepam is initially used because of its rapid onset of action, consider changing to oral baclofen if long-term treatment is indicated.

Give oral diazepam treatment as a bedtime dose. If the response is unsatisfactory consider:

- Increasing the dose or
- Adding a daytime dose

Start oral baclofen treatment with a low dose and increase the dose stepwise over about 4 weeks to achieve the optimum therapeutic effect.

Continue using oral diazepam or oral baclofen if they have a clinical benefit and are well tolerated, but think about stopping the treatment whenever the child or young person's management programme is reviewed and at least every 6 months.

If adverse effects (such as drowsiness) occur with oral diazepam or oral baclofen, think about reducing the dose or stopping treatment.

If the response to oral diazepam and oral baclofen used individually for 4–6 weeks is unsatisfactory, consider a trial of combined treatment using both drugs.

If a child or young person has been receiving oral diazepam and/or baclofen for several weeks, ensure that when stopping these drugs the dose is reduced in stages to avoid withdrawal symptoms.

In children and young people with spasticity in whom dystonia is considered to contribute significantly to problems with posture, function, and pain, consider a trial of oral drug treatment, for example with trihexyphenidyl¹, levodopa² or baclofen³.

Botulinum Toxin Type A

General Principles

Consider botulinum toxin type A⁴ treatment in children and young people in whom focal spasticity of the upper limb is:

- Impeding fine motor function
- Compromising care and hygiene
- Causing pain
- Impeding tolerance of other treatments, such as orthoses
- Causing cosmetic concerns to the child or young person

Consider botulinum toxin type A⁴ treatment where focal spasticity of the lower limb is:

- Impeding gross motor function
- Compromising care and hygiene
- Causing pain
- Disturbing sleep
- Impeding tolerance of other treatments, such as orthoses and use of equipment to support posture
- Causing cosmetic concerns to the child or young person

Consider botulinum toxin type A⁴ treatment after an acquired non-progressive brain injury if rapid-onset spasticity is causing postural or functional difficulties.

Consider a trial of botulinum toxin type A⁵ treatment in children and young people with spasticity in whom focal dystonia is causing serious problems, such as postural or functional difficulties or pain.

Do not offer botulinum toxin type A treatment if the child or young person:

- Has severe muscle weakness
- Had a previous adverse reaction or allergy to botulinum toxin type A
- Is receiving aminoglycoside treatment

Be cautious when considering botulinum toxin type A treatment if:

- The child or young person has any of the following:
 - A bleeding disorder, for example due to anti-coagulant therapy
 - Generalised spasticity
 - Fixed muscle contractures
 - Marked bony deformity or
- There are concerns about the child or young person's likelihood of engaging with the post-treatment adapted physical therapy programme (see the related recommendation in the section on Physical Therapy; Specific Strategies, above).

When considering botulinum toxin type A treatment, perform a careful assessment of muscle tone, range of movement, and motor function to:

- Inform the decision as to whether the treatment is appropriate
- Provide a baseline against which the response to treatment can be measured

A physiotherapist or an occupational therapist should be involved in the assessment.

When considering botulinum toxin type A treatment, give the child or young person and their parents or carers information about:

- The possible benefits and the likelihood of achieving the treatment goals
- What the treatment entails, including:
 - The need for assessments before and after the treatment
 - The need to inject the drug into the affected muscles
 - The possible need for repeat injections
 - The benefits, where necessary, of analgesia, sedation, or general anaesthesia
 - The need to use serial casting or an orthosis after the treatment in some cases
- Possible important adverse effects (see also the recommendation in this section, under "Delivering Treatment," below).

Botulinum toxin type A treatment (including assessment and administration) should be provided by healthcare professionals within the network team who have expertise in child neurology and musculoskeletal anatomy.

Delivering Treatment

Before starting treatment with botulinum toxin type A, tell children and young people and their parents or carers:

- To be aware of the following rare but serious complications of botulinum toxin type A treatment:
 - Swallowing difficulties
 - Breathing difficulties
- How to recognise signs suggesting these complications are present
- That these complications may occur at any time during the first week after the treatment and
- That if these complications occur the child or young person should return to hospital immediately

To avoid distress to the child or young person undergoing treatment with botulinum toxin type A, think about the need for:

- Topical or systemic analgesia or anaesthesia
- Sedation (see the NICE guideline Sedation in children and young people, [NICE clinical guideline 112])

Consider ultrasound or electrical muscle stimulation to guide the injection of botulinum toxin type A.

Consider injecting botulinum toxin type A into more than one muscle if this is appropriate to the treatment goal, but ensure that maximum dosages are not exceeded.

After treatment with botulinum toxin type A, consider an orthosis to:

- Enhance stretching of the temporarily weakened muscle and
- Enable the child or young person to practice functional skills

If an orthosis is indicated after botulinum toxin type A, but limited passive range of movement would make this difficult, consider first using serial casting to stretch the muscle. To improve the child or young person's ability to tolerate the cast, and to improve muscle stretching, delay casting until 2–4 weeks after the botulinum toxin type A treatment.

Ensure that children and young people who receive treatment with botulinum toxin type A are offered timely access to orthotic services.

Continuing Assessment

Perform an assessment of muscle tone, range of movement, and motor function:

- 6–12 weeks after injections to assess the response
 - 12–26 weeks after injections to inform decisions about further injections
- These assessments should preferably be performed by the same healthcare professionals who undertook the baseline assessment.

Consider repeat injections of botulinum toxin type A if:

- The response in relation to the child or young person's treatment goal was satisfactory, and the treatment effect has worn off
- New goals amenable to this treatment are identified

Intrathecal Baclofen

General Principles

Consider treatment with continuous pump-administered intrathecal baclofen⁶ in children and young people with spasticity if, despite the use of non-invasive treatments, spasticity or dystonia are causing difficulties with any of the following:

- Pain or muscle spasms
- Posture or function
- Self-care (or ease of care by parents or carers)

Be aware that children and young people who benefit from continuous pump-administered intrathecal baclofen typically have:

- Moderate or severe motor function problems (GMFCS level III, IV, or V)

- Bilateral spasticity affecting upper and lower limbs

Be aware of the following contraindications to treatment with continuous pump-administered intrathecal baclofen:

- The child or young person is too small to accommodate an infusion pump
- Local or systemic intercurrent infection

Be aware of the following potential contraindications to treatment with continuous pump-administered intrathecal baclofen:

- Co-existing medical conditions (for example, uncontrolled epilepsy or coagulation disorders)
- A previous spinal fusion procedure
- Malnutrition, which increases the risk of post-surgical complications (for example, infection or delayed healing)
- Respiratory disorders with a risk of respiratory failure

If continuous pump-administered intrathecal baclofen is indicated in a child or young person with spasticity in whom a spinal fusion procedure is likely to be necessary for scoliosis, implant the infusion pump before performing the spinal fusion.

When considering continuous pump-administered intrathecal baclofen, balance the benefits of reducing spasticity against the risk of doing so because spasticity sometimes supports function (for example, by compensating for muscle weakness). Discuss these possible adverse effects with the child or young person and their parents or carers.

When considering continuous pump-administered intrathecal baclofen, inform children and young people and their parents or carers verbally and in writing (or appropriate formats) about:

- The surgical procedure used to implant the pump
- The need for regular hospital follow-up visits
- The requirements for pump maintenance
- The risks associated with pump implantation, pump-related complications, and adverse effects that might be associated with intrathecal baclofen infusion.

Intrathecal Baclofen Testing

Before making the final decision to implant the intrathecal baclofen pump, perform an intrathecal baclofen test to assess the therapeutic effect and to check for adverse effects.

Before intrathecal baclofen testing, inform children and young people and their parents or carers verbally and in writing (or appropriate formats) about:

- What the test will entail
- Adverse effects that might occur with testing
- How the test might help to indicate the response to treatment with continuous pump-administered intrathecal baclofen, including whether:
 - The treatment goals are likely to be achieved
 - Adverse effects might occur

Before performing the intrathecal baclofen test, assess the following where relevant to the treatment goals:

- Spasticity
 - Dystonia
 - The presence of pain or muscle spasms
 - Postural difficulties, including head control
 - Functional difficulties
 - Difficulties with self-care (or ease of care by parents or carers)
- If necessary, assess passive range of movement under general anaesthesia.

The test dose or doses of intrathecal baclofen should be administered using a catheter inserted under general anaesthesia.

Assess the response to intrathecal baclofen testing within 3–5 hours of administration. If the child or young person is still sedated from the general anaesthetic at this point, repeat the assessment later when they have recovered.

When deciding whether the response to intrathecal baclofen is satisfactory, assess the following where relevant to the treatment goals:

- Reduction in spasticity
- Reduction in dystonia
- Reduction in pain or muscle spasms
- Improved posture, including head control
- Improved function
- Improved self-care (or ease of care by parents or carers)

Discuss with the child or young person and their parents or carers their views on the response to the intrathecal baclofen test. This should include their assessment of the effect on self-care (or ease of care by parents or carers). Consider using a standardised questionnaire to document their feedback.

Intrathecal baclofen testing should be:

- Performed in a specialist neurosurgical centre within the network that has the expertise to carry out the necessary assessments
- Undertaken in an inpatient setting to support a reliable process for assessing safety and effectiveness

Initial and post-test assessments should be performed by the same healthcare professionals in the specialist neurosurgical centre.

Continuous Pump-Administered Intrathecal Baclofen

Before implanting the intrathecal baclofen pump, inform children and young people and their parents or carers, verbally and in writing (or appropriate formats), about:

- Safe and effective management of continuous pump-administered intrathecal baclofen
- The effects of intrathecal baclofen, possible adverse effects, and symptoms and signs suggesting the dose is too low or too high
- The potential for pump-related complications
- The danger of stopping the continuous pump-administered intrathecal baclofen infusion suddenly
- The need to attend hospital for follow-up appointments, for example to refill and reprogram the infusion pump
- The importance of seeking advice from a healthcare professional with expertise in intrathecal baclofen before stopping the treatment

Implant the infusion pump and start treatment with continuous pump-administered intrathecal baclofen within 3 months of a satisfactory response to intrathecal baclofen testing (see the related recommendation in the section on Intrathecal Baclofen Testing, above).

Support children and young people receiving treatment with continuous pump-administered intrathecal baclofen and their parents or carers by offering regular follow-up with the network team, and a consistent point of contact with the specialist neurosurgical centre.

Monitor the response to continuous pump-administered intrathecal baclofen. This monitoring should preferably be performed by the healthcare professionals in the regional specialist neurosurgical centre who performed the pre-implantation assessments.

When deciding whether the response to continuous pump-administered intrathecal baclofen is satisfactory, assess the following where relevant to the treatment goals:

- Reduction in spasticity
- Reduction in dystonia
- Reduction in pain or muscle spasms
- Improved posture, including head control
- Improved function
- Improved self-care (or ease of care by parents or carers)

Titrate the dose of intrathecal baclofen after pump implantation, if necessary, to optimise effectiveness.

If treatment with continuous pump-administered intrathecal baclofen does not result in a satisfactory response (see the related recommendation in this section, above), check that there are no technical faults in the delivery system and that the catheter is correctly placed to deliver the drug to the intrathecal space. If no such problems are identified, consider reducing the dose gradually to determine whether spasticity and associated symptoms increase.

If continuous pump-administered intrathecal baclofen therapy is unsatisfactory, the specialist neurosurgical centre and other members of the network team should discuss removing the pump and alternative management options with the child or young person and their parents or carers.

As the infusion pump approaches the end of its expected lifespan, consider reducing the dose gradually to enable the child or young person and

their parents or carers to decide whether or not to have a new pump implanted.

Orthopaedic Surgery

Consider orthopaedic surgery as an important adjunct to other interventions in the management programme for some children and young people with spasticity. Timely surgery can prevent deterioration and improve function.

An assessment should be performed by an orthopaedic surgeon within the network team if:

- Based on clinical findings (see the related recommendation in the section on Principles of Care; Monitoring, above) or radiological monitoring, there is concern that the hip may be displaced
- Based on clinical or radiological findings there is concern about spinal deformity

Consider an assessment by an orthopaedic surgeon in the network team for children and young people with:

- Hip migration greater than 30% or
- Hip migration percentage increasing by more than 10 percentage points per year

Consider an assessment by an orthopaedic surgeon in the network team if any of the following are present:

- Limb function is limited (for example, in walking or getting dressed) by unfavourable posture or pain, as a result of muscle shortening, contractures, or bony deformities
- Contractures of the shoulder, elbow, wrist, or hand cause difficulty with skin hygiene
- The cosmetic appearance of the upper limb causes significant concern for the child or young person

Before undertaking orthopaedic surgery, the network team should discuss and agree with the child or young person and their parents or carers:

- The possible goals of surgery and the likelihood of achieving them
- What the surgery will entail, including any specific risks
- The rehabilitation programme, including:
 - How and where it will be delivered
 - What the components will be, for example a programme of adapted physical therapy, the use of orthoses, oral drugs, or botulinum toxin type A

Orthopaedic surgery should:

- Be undertaken by surgeons in the network team who are expert in the concepts and techniques involved in surgery for this group of patients and
- Take place in a paediatric setting

The decision to perform orthopaedic surgery to improve gait should be informed by a thorough pre-operative functional assessment, preferably including gait analysis.

If a child or young person will need several surgical procedures at different anatomical sites to improve their gait, perform them together if possible (single-event multilevel surgery), rather than individually over a period of time.

Assess the outcome of orthopaedic surgery undertaken to improve gait 1–2 years later. By then full recovery may be expected and the outcome of the procedure can be more accurately determined.

Selective Dorsal Rhizotomy

Consider selective dorsal rhizotomy to improve walking ability in children and young people with spasticity at GMFCS level II or III:

- Patient selection and treatment should be carried out by a multidisciplinary team with specialist training and expertise in the care of spasticity, and with access to the full range of treatment options.
- Discuss the irreversibility of the treatment, the known complications and the uncertainties over long-term outcomes with children and young people, and their parents and/or carers (see also *Selective dorsal rhizotomy for spasticity in cerebral palsy*, [NICE interventional procedure guidance 373](#)).
- Teams offering selective dorsal rhizotomy should participate in a coordinated national agreed programme to collect information on short- and long-term outcomes on all patients assessed for selective dorsal rhizotomy, whether or not selective dorsal rhizotomy is performed.

These recorded outcomes should include measures of muscle tone, gross motor function, neurological impairment, spinal deformity, quality of life, and need for additional operations, with nationally agreed consistent definitions.

Notes

¹At the time of publication (July 2012), trihexyphenidyl did not have United Kingdom (UK) marketing authorisation for use in the treatment of dystonia associated with spasticity, and its use is not recommended in children. However, it is used in the UK for the treatment of dystonia in children and young people with spasticity. Informed consent should be obtained and documented.

²At the time of publication (July 2012), levodopa (which is always marketed in combination with an extra-cerebral dopa-decarboxylase inhibitor) did not have UK marketing authorisation for use in the treatment of dystonia associated with spasticity, and its use is not recommended in children or young people. However, it is used in the UK for the treatment of dystonia in children and young people with spasticity. Informed consent should be obtained and documented.

³At the time of publication (July 2012), baclofen did not have UK marketing authorisation for use in the treatment of dystonia associated with spasticity. However, it is used in the UK for the treatment of dystonia in children and young people with spasticity. Informed consent should be obtained and documented.

⁴At the time of publication (July 2012), some botulinum toxin type A products had UK marketing authorisation for use in the treatment of focal spasticity in children, young people and adults, including the treatment of dynamic equinus foot deformity due to spasticity in ambulant paediatric cerebral palsy patients, 2 years of age or older. Other products had UK marketing authorisation only for use on the face in adults or for post-stroke spasticity of the upper limb in adults. Botulinum toxin units are not interchangeable from one product to another. Details of licensed indications and doses for individual products are available at the electronic Medicines Compendium. Where appropriate, informed consent should be obtained and documented.

⁵At the time of publication (July 2012), botulinum toxin type A did not have UK marketing authorisation for use in the treatment of focal dystonia associated with spasticity. However, it is used in the UK for the treatment of dystonia in children and young people with spasticity. Informed consent should be obtained and documented.

⁶At the time of publication (July 2012), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years, nor did it have UK marketing authorisation for use in the treatment of dystonia associated with spasticity. Where appropriate, informed consent should be obtained and documented.

Clinical Algorithm(s)

A NICE Pathway on spasticity in children and young people is provided at the [National Institute for Health and Clinical Excellence \(NICE\) Web site](#) . Care pathways are also provided in the full version of the original guideline document (see the "Availability of Companion Documents" field).

Scope

Disease/Condition(s)

Spasticity and co-existing motor disorders and their early musculoskeletal complications, including spasticity from cerebral palsy

Guideline Category

Counseling

Evaluation

Management

Prevention

Treatment

Clinical Specialty

Family Practice

Internal Medicine

Neurological Surgery

Neurology

Nursing

Orthopedic Surgery

Pediatrics

Physical Medicine and Rehabilitation

Surgery

Intended Users

Advanced Practice Nurses

Allied Health Personnel

Health Care Providers

Nurses

Occupational Therapists

Patients

Physical Therapists

Physician Assistants

Physicians

Psychologists/Non-physician Behavioral Health Clinicians

Public Health Departments

Social Workers

Guideline Objective(s)

- To provide a clinical guideline on the management of spasticity in children and young people with a non-progressive brain disorder
- To help healthcare professionals to select and use appropriate treatments for individual children and young people

Target Population

Children and young people from birth up to their 19th birthday who have spasticity as a result of a non-progressive brain disorder, including those with spasticity resulting from cerebral palsy and those with spasticity resulting from a non-progressive brain injury acquired later in childhood or adolescence

Note: These guidelines are not intended for use in the following patients:

Adults 19 years and older

Children and young people with spasticity resulting from a progressive brain disorder. However, many of the recommendations on the management of spasticity might also apply to these children and young people.

Children with a pure dystonia or other motor disorders which do not co-exist with spasticity

Interventions and Practices Considered

1. Providing access to a team of healthcare professionals experienced in the care of children and young people with spasticity
2. Offering an individualised, goal-focused management programme that is developed and implemented in partnership with the child or young

- person and their parents or carers
3. Supporting the child or young person and their parents or carers
 4. Monitoring for response to treatments, worsening of spasticity, developing secondary consequences of spasticity, and need to change their individualised goals
 5. Monitoring for risk of hip displacement, including hip x-ray
 6. Individualised physical therapy (physiotherapy and/or occupational therapy) programme
 7. Orthoses
 8. Oral diazepam, baclofen, trihexyphenidyl or levodopa
 9. Botulinum toxin type A injection
 10. Continuing assessment of muscle tone, range of movement, and motor function
 11. Continuous pump-administered intrathecal baclofen
 12. Intrathecal baclofen test to assess the therapeutic effect and to check for adverse effects
 13. Orthopaedic surgery
 14. Selective dorsal rhizotomy
 15. Advising patients and carers about adverse events of treatment

Major Outcomes Considered

- Reduction of spasticity/dystonia
- Optimisation of movement and function
- Reduction of pain
- Adverse effects of interventions
- Acceptability and tolerability in children and young people
- Health related quality of life
- Cost-effectiveness
- Quality-adjusted life years (QALYs)

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Collaborating Centre for Women's and Children's Health (NCGC) on behalf of the National Institute for Health and Clinical Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

Developing Review Questions and Protocols and Identifying Evidence

The Guideline Development Group (GDG) formulated review questions based on the scope and prepared a protocol for each review question (see Appendix D of the full version of the guideline; see the "Availability of Companion Documents" field). These formed the starting point for systematic reviews of relevant evidence.

Specific outcomes considered during the evaluation of published evidence are outlined in Appendix E of the full version of the guideline. Published evidence was identified by applying systematic search strategies (see Appendix F of the full version of the guideline) to the following databases: Medline, Medline In-Process, EMBASE, Cumulative Index to Nursing and Allied Health Literature (CINAHL), and three Cochrane databases (Cochrane Central Register of Controlled Trials, Cochrane Database of Systematic Reviews, and the Database of Abstracts of Reviews of Effects). Searches to identify economic studies were undertaken using Medline, EMBASE, the Cochrane Central Register of Controlled Trials, the NHS Economic Evaluation Database (NHS EED), and the Health Technology Assessment (HTA) database.

Dates of searching and database coverage are given with the details of the search strategies in Appendix F of the full version of the guideline.

Where appropriate, review questions were grouped together for searching. The search strategies from National Institute for Health and Clinical Excellence (NICE) interventional procedure guidance (IPG) [Selective dorsal rhizotomy for spasticity in cerebral palsy](#) (NICE IPG 373, 2010) were used for the selective dorsal rhizotomy (SDR) review. The search for the physical therapy review was limited by date (the search was limited to articles published after 1970), but the remaining searches were not. Animal studies were excluded from Medline and both Medline and EMBASE were limited to English-language studies only. Studies conducted in adult populations were not excluded using search filters. Scottish Intercollegiate Guidelines Network (SIGN) search filters were used to identify particular study designs, such as randomised controlled trials. There was no systematic attempt to search grey literature (conference abstracts, theses, or unpublished trials), nor was hand searching of journals not indexed on the databases undertaken.

Towards the end of the guideline development process, the searches were updated and re-executed to include evidence published and indexed in the databases before 8 August 2011.

Incorporating Health Economics

The GDG prioritised a number of review questions where it was thought that economic considerations would be particularly important in formulating recommendations. Systematic searches for published economic evidence were undertaken for these questions.

Number of Source Documents

Number of papers included:

- Question 1: Clinical search: 14; Economic search: 0
- Question 2: Clinical search: 6; Economic search: 0
- Question 3: Clinical search: 9; Economic search: 0
- Question 4: Clinical search: 9; Economic search: 3
- Questions 5 and 6: Clinical search: 10; Economic search: 0
- Questions 7 and 8: Clinical search: 4; Economic search: 0
- Question 9: Clinical Search: 7

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Overall Quality of Outcome Evidence in GRADE (Grading of Recommendations Assessment, Development and Evaluation)

Level	Description
High	Further research is very unlikely to change confidence in the estimate of effect
Moderate	Further research is likely to have an important impact on confidence in the estimate of effect and may change the estimate
Low	Further research is very likely to have an important impact on confidence in the estimate of effect and is likely to change the estimate
Very Low	Any estimate of effect is very uncertain

Methods Used to Analyze the Evidence

Meta-Analysis

Description of the Methods Used to Analyze the Evidence

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Collaborating Centre for Women's and Children's Health (NCGC) on behalf of the National Institute for Health and Clinical Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

Reviewing and Synthesising Evidence

Evidence relating to clinical effectiveness was reviewed and synthesised according to the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach (see the "Rating Scheme for the Strength of the Evidence" field). In this approach, the quality of the evidence identified for each outcome listed in the review protocol is assessed according to the factors listed below and an overall quality rating (very low, low, moderate, or high) is assigned by combining ratings for the individual factors.

- Study design (as an indicator of intrinsic bias; this determines the initial quality rating)
- Limitations in the design or execution of the study (including concealment of allocation, blinding, loss to follow up; these and other sources of bias can reduce the quality rating)
- Inconsistency of effects across studies (this can reduce the quality rating where more than one study is considered)
- Indirectness (the extent to which the available evidence fails to address the specific review question; this can reduce the quality rating)
- Imprecision (the extent to which the point estimate or its confidence interval (CI) reflects a clinically important difference; this can reduce the quality rating)
- Other considerations (including large magnitude of effect, evidence of a dose–response relationship or confounding variables likely to have reduced the magnitude of an effect; these can increase the quality rating in observational studies, provided no downgrading for other features has occurred).

The Guideline Development Group (GDG) considered that reduction of spasticity alone without concomitant clinically meaningful improvement in other patient-centred outcomes would be insufficient to recommend an intervention. At the start of the guideline development period, the GDG discussed, specified and prioritised units of measurement for each main outcome detailed in the scope. As far as possible the GDG selected similar units derived from validated and clinically used assessment techniques to be applied across each review for consistency (see Appendix E of the full version of the guideline; see the "Availability of Companion Documents" field). Where outcomes from validated assessment techniques were not available in the literature, outcomes from non-validated tools were discussed with GDG members and included only on their advice.

The type of review question determines the highest level of evidence that may be sought. For issues of treatment, the highest possible evidence level is a well conducted systematic review or meta-analysis of randomised controlled trials (RCTs), or an individual RCT. In the GRADE approach, a body of evidence based entirely on such studies has an initial quality rating of high, and this may be downgraded to moderate, low, or very low if factors listed above are not addressed adequately.

Various approaches may be used to assess imprecision in the GRADE framework. One such approach is to downgrade for imprecision on the basis of inadequate event rates (fewer than 300 for dichotomous outcomes) or inadequate study population size (less than 400 participants for continuous outcomes). No outcomes in this guideline met these criteria; therefore, while footnotes were made to this effect, the outcomes were not downgraded based on these criteria. For dichotomous outcomes, where a 95% CI for a relative risk (RR) or odds ratio (OR) crossed the line of no effect and either one or both of the GRADE default lower or upper thresholds for downgrading (0.75 or 1.25), imprecision was rated as serious. Where the 95% CI was entirely below 0.75 or entirely above 1.25, or entirely between 0.75 and 1.25, the outcome was not downgraded for imprecision and the result could be interpreted as being clinically important.

The results of many different assessment tools were examined as continuous outcomes in this guideline. The GDG sought to identify clinically important differences for the outcomes of each assessment tool. Where possible, the GDG's definitions were applied to data extracted from published articles to inform decisions about whether or not the quality of the evidence should be downgraded for imprecision. Where the GDG was unable to specify a clinically important difference, or the data were insufficient to permit extrapolation, the outcome was downgraded. Further details of the GDG's considerations with regard to defining clinically important differences for continuous outcome measures prioritised for inclusion in the guideline (such as scores from various assessment tools) are summarised in Appendix E of the full version of the guideline.

For each review question the highest available level of evidence was sought. Where appropriate, for example, if a systematic review, meta-analysis or RCT was identified to answer a question directly, studies of a weaker design were not considered. Where systematic reviews, meta-analyses

and RCTs were not identified, other appropriate experimental or observational studies were included following discussion with the GDG.

Some studies were excluded from the guideline reviews after obtaining copies of the corresponding publications because they did not meet inclusion criteria specified by the GDG and recorded in the review protocols (see Appendix H of the full version of the guideline). The characteristics of each included study were summarised in evidence tables for each review question (see Appendix I of the full version of the guideline). Where possible, dichotomous outcomes were presented as RRs or ORs with 95% CIs, and continuous outcomes were presented as mean differences (MDs) with 95% CIs or standard deviations (SDs).

The body of evidence identified for each review question (or part of a review question) was presented in the form of a GRADE evidence profile summarising the quality of the evidence and the findings (pooled relative and absolute effect sizes and associated CIs). Where possible, the body of evidence corresponding to each outcome specified in the review protocol was subjected to quantitative meta-analysis. In such cases, pooled effect sizes were presented as pooled RRs, pooled ORs or weighted mean differences (WMDs). By default, meta-analyses conducted specifically for the guideline (see Tables 7.5 and 10.2 of the full version of the guideline) used a fixed effect model, and where statistically significant heterogeneity was identified a random effects model was used. Forest plots for all meta-analyses conducted specifically for the guideline are presented in Appendix J of the full version of the guideline. The meta-analyses presented in Tables 7.1, 7.3 and 7.6 of the full version of the guideline were reported in a Cochrane systematic review. Some of these meta-analyses were conducted using a fixed effect model and others were conducted using a random effects model. Where statistically significant heterogeneity was identified, the guideline evidence statements report findings from the individual studies that contributed to the meta-analysis. GRADE findings are presented in full in Appendix K of the full version of the guideline and abbreviated versions (summary of findings without the individual components of the quality assessment) are presented in the full version of the guideline.

Specific Considerations for This Guideline

Physiotherapy and occupational therapy are core treatments for children and young people with spasticity although their primary aim is not to reduce spasticity (this is also true of orthoses). The GDG acknowledged that most children and young people included in clinical research studies would have received physiotherapy and/or occupational therapy. The GDG agreed to use the term 'physical therapy' to encompass all interventions that would normally be prescribed or performed by a physiotherapist or an occupational therapist. When direct reference was made to a particular study, however, the terms used by the authors of the study publications were used. Physiotherapists and occupational therapists are referred to collectively as physical therapists in this guideline. In the publications reviewed for the guideline it was not always clear exactly what form of physical therapy had been delivered, what sort of healthcare professional had prescribed or administered the physical therapy, how frequently or intensively the physical therapy had been administered, or whether the intervention and comparison groups had received the same forms of physical therapy. Nevertheless, those details of physical therapy interventions that were reported in each included study were recorded in the corresponding evidence tables.

With regard to the age of participants in the research studies reviewed for the guideline, the term child is used to refer to people under the age of 11 years, and young person is used to refer to those aged 11–19 years.

Incorporating Health Economics

For economic evaluations, no standard system of grading the quality of evidence exists and included papers were assessed using a quality assessment checklist based on good practice in economic evaluation. Reviews of the (very limited) relevant published health economic literature are presented alongside the corresponding clinical effectiveness reviews.

Health economic considerations were aided by original economic analysis undertaken as part of the development process. For this guideline the areas prioritised for economic analysis were as follows:

- Physical therapy (physiotherapy and/or occupational therapy)
- Orthoses
- Botulinum toxin
- Continuous pump-administered intrathecal baclofen
- Orthopaedic surgery
- Selective dorsal rhizotomy

Details of the health economic analyses conducted for the guideline are presented in Chapter 11 of the full version of the guideline.

The GDG considered using the EuroQol Group's EQ-5D instrument to evaluate quality of life but had reservations about its application in children and young people. None of the studies identified for inclusion in the guideline reviews used the EQ-5D for children and there was insufficient clinical evidence available for translation into the EQ-5D for children, or for subsequent health economic interpretation or analysis.

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Collaborating Centre for Women's and Children's Health (NCGC) on behalf of the National Institute for Health and Clinical Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

This guidance was developed in accordance with the NICE guideline development process outlined in the 2009 edition of The Guidelines Manual (see the "Availability of Companion Documents" field).

The NCGC established a Guideline Development Group (GDG), which reviewed the evidence and developed the recommendations. An independent Guideline Review Panel oversaw the development of the guideline.

Organisations with interests in the management of spasticity, co-existing motor disorders and their early musculoskeletal complications in children and young people with non-progressive brain disorders were encouraged to register as stakeholders for the guideline. Registered stakeholders were consulted throughout the guideline development process.

In accordance with NICE's Equality Scheme, ethnic and cultural considerations and factors relating to disabilities have been considered by the GDG throughout the development process and specifically addressed in individual recommendations where relevant. Further information is detailed in [NICE's Equality Scheme](#) .

Evidence to Recommendations

For each review question recommendations for clinical care were derived using, and linked explicitly to, the evidence that supported them. In the first instance, short clinical and, where appropriate, cost effectiveness evidence statements were drafted by the technical team and presented alongside the evidence profiles to be agreed by the GDG. Statements summarising the GDG's interpretation of the evidence and any extrapolation from the evidence used to form recommendations were also prepared to ensure transparency in the decision-making process. The criteria used in moving from evidence to recommendations were as follows:

- Relative value placed on the outcomes considered
- Consideration of the clinical benefits and harms
- Consideration of net health benefits and resource use
- Quality of the evidence
- Other considerations (including equalities issues)

In areas where no substantial clinical evidence was identified, the GDG members considered other evidence-based guidelines and consensus statements or used their collective experience to identify good practice. The health economics justification in areas of the guideline where the use of National Health Service (NHS) resources (interventions) was considered was based on GDG consensus with regard to the likely cost effectiveness implications of the recommendations. The GDG members also identified areas where evidence to answer their review questions was lacking and used this information to formulate recommendations for future research.

Towards the end of the guideline development process, formal consensus methods incorporating anonymous voting were used to consider all the clinical care recommendations and research recommendations that had previously been drafted. The GDG identified nine key priorities for implementation (key recommendations) and five high priority (key) research recommendations. The key priorities for implementation were those recommendations thought likely to have the biggest impact on clinical care and outcomes in the NHS as a whole. The key research recommendations were selected in a similar way.

Rating Scheme for the Strength of the Recommendations

Not applicable

Cost Analysis

Where possible, economic analysis has been developed by working backwards from the National Institute of Health and Clinical Excellence (NICE) cost-effectiveness threshold to determine what level of effectiveness would be necessary in order to find an intervention cost effective. This type of analysis does not give cost effectiveness results, but provides a framework within which to decide whether a treatment is likely to be good value in terms of National Health Service (NHS) resources.

For almost all the interventions considered in the guideline, published evidence of cost-effectiveness was completely lacking. In the following areas, further analysis was undertaken to support the GDG's decision making:

- Physical therapy versus no active treatment
- Ankle-foot orthoses versus no active treatment
- Botulinum toxin versus oral drugs in combination with other interventions
- Continuous pump-administered intrathecal baclofen (CITB), including intrathecal baclofen (ITB) testing before CITB versus no ITB testing, and CITB versus oral drugs in combination with other interventions
- Orthopaedic surgery versus no active treatment
- Selective dorsal rhizotomy versus no active treatment

None of the analyses presented in the Health Economics chapter of the full version of the original guideline document follow NICE's reference case for health economic analysis because of the lack of evidence for effectiveness and because the GDG was not able to quantify the benefits of treatment in a way that could be used in an economic analysis using consensus values for unknown parameters.

In all of the topics considered for economic evaluation resource use and costs were quantified. Details of the methods used in relation to each review question are presented in Chapter 11 of the full version of the original guideline document. For each question the following are reported: review of published economic literature; description of resource use and costs; and conclusions of the analysis. A discussion of results was also included for questions for which a full health economic analysis was undertaken.

For many of the treatments considered in the guideline, the Guideline Development Group (GDG) felt that the benefits to health-related quality of life outweighed the potential harms. Patient selection is important, particularly for the ITB pump, orthopaedic surgery, and selective dorsal rhizotomy, as only certain groups of patients are likely to benefit and treatment will not be appropriate for other groups. Patient choice is also important as their active participation, such as in physical therapy programmes and use of orthoses, is key to the success of several treatments.

For each review question considered, the full version of the original guideline document includes a health economic summary based on evidence and GDG opinion.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

Registered stakeholder organisations were invited to comment on the draft scope and the draft guideline. Stakeholder organisations were also invited to undertake a prepublication check of the final guideline to identify factual inaccuracies. The Guideline Development Group (GDG) carefully considered and responded to all comments received from stakeholder organisations. The comments and responses, which were reviewed independently for National Institute for Health and Clinical Excellence (NICE) by a Guidelines Review Panel, are published on the NICE website.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of evidence supporting the recommendations is not specifically stated.

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate management of spasticity and co-existing motor disorders and their early musculoskeletal complications in children and young people with nonprogressive brain disorders

Refer to the "Trade off between clinical benefits and harms" sections in the full version of the guideline (see the "Availability of Companion Documents" field) for the specific for individual recommendations.

Potential Harms

- The Guideline Development Group (GDG) agreed that adverse events associated with physical therapy were likely to be relatively uncommon and often minor (for example minor injury, discomfort or pain) or manageable with modification of the physical therapy programme.
- The GDG acknowledged that intensive physical therapy could be associated with significant disruption to the lives of the child or young person and their parents or carers, but thought that this potential risk could be mitigated through recommendations to ensure that these individuals should be provided with adequate information to allow them to make informed choices about the nature of the physical therapy programme being undertaken.
- Orthoses may be beneficial in terms of enhancing function and posture, but they may have disadvantages too. They may be considered to be unsightly or cause discomfort and pressure injuries and, if used inappropriately, they may affect function adversely. In the GDG's experience, adverse effects were deemed to have a major impact on the child or young person's ability and willingness to accept or tolerate an orthosis. There was also consensus in the group that discomfort, skin injury, sleep disturbance and so on are more likely to occur if orthoses are badly designed, ill-fitting or worn, but that adverse effects should mainly be preventable with careful design and fitting of the devices. A longer-term risk of muscle wasting and weakness resulting from immobilisation with orthoses was identified.
- One randomised controlled trial reported that bedtime administration of diazepam was not associated with daytime drowsiness. However, the GDG noted that the dose of diazepam employed in the study was less than that usually used in United Kingdom practice and recommended in the summary of product characteristics. The GDG considered that with higher doses the outcomes might have been different, and the likelihood of sedation and increased oral secretions (a recognised side effect of diazepam treatment in children and young people with spasticity) might have been greater.
- Drowsiness was reported as a specific side effect of baclofen and this appeared to be dose related.
- The GDG considered that there were potential adverse effects associated with withdrawal of diazepam and baclofen after a long period of treatment. The group therefore recommended that discontinuation after several weeks' use should be accomplished through staged reductions in dose to avoid withdrawal symptoms.
- The GDG considered adverse effects of botulinum toxin.
 - Four serious adverse events (requiring hospitalisation) were reported for use of botulinum toxin in one upper limb randomised controlled trial within the Cochrane systematic review and meta-analysis considered for the guideline. These all occurred in children and young people known to have co-existing medical conditions. Severe adverse events of concern, but not reported in the evidence reviewed, included swallowing and breathing difficulties following injection around the shoulder, neck, and thorax. Other reported adverse effects included short-term muscle weakness and less specific complaints.
 - In the lower limb, reported adverse events included pain after the botulinum toxin type A injection, increased frequency of falls, incontinence, short-term muscle weakness, and other less specific complaints. The GDG felt that these side effects were important to note when seeking consent for the procedure, but noted that they are infrequently reported and usually short lived.
- In general, the GDG believed that adverse effects associated with intrathecal baclofen testing would occur only occasionally and the effects would usually be minor.
- The GDG recognised, based on the evidence and their clinical consensus, that there were potential risks associated with the continuous pump-administered intrathecal baclofen treatment and these included all the general risks associated with surgery, such as the need for general anaesthetic.
- The GDG members noted a number of risks associated with orthopaedic surgery.
 - The risks of orthopaedic surgery for the management of spasticity included all the general risks of any surgical procedure, such as the need for general anaesthesia.
 - It might take 1–2 years for patients to recover fully and gain the full benefit from single-event multilevel surgery. Even if surgery might be beneficial in principle, the tendency for spasticity and its complications to progress over time might hide such benefits.
 - Orthopaedic surgery may be a major procedure with attendant risks of pain, haemorrhage, and infection, but immediate post-

operative pain after surgery in the lower limbs could be managed effectively with epidural analgesia.

- The risks of haemorrhage requiring blood transfusion would vary, but they become greater with more extensive surgery.
- In operations that require division and stabilisation of bone with metallic internal fixation devices there is a risk of non-union of bone.
- The GDG was aware of the adverse effects associated with X-rays (used to monitor for hip problems following orthopaedic surgery).
- Although the risks of permanent morbidity following selective dorsal rhizotomy surgery are low, the potential consequences are serious.

Refer to the "Trade off between clinical benefits and harms" sections in the full version of the guideline for specific recommendations.

Contraindications

Contraindications

- Do not offer botulinum toxin type A treatment if the child or young person:
 - Has severe muscle weakness
 - Had a previous adverse reaction or allergy to botulinum toxin type A
 - Is receiving aminoglycoside treatment
- The following are contraindications to treatment with continuous pump-administered intrathecal baclofen:
 - The child or young person is too small to accommodate an infusion pump
 - Local or systemic intercurrent infection
- The following are potential contraindications to treatment with continuous pump-administered intrathecal baclofen:
 - Co-existing medical conditions (for example, uncontrolled epilepsy or coagulation disorders)
 - A previous spinal fusion procedure
 - Malnutrition, which increases the risk of post-surgical complications (for example, infection or delayed healing)
 - Respiratory disorders with a risk of respiratory failure

Qualifying Statements

Qualifying Statements

- This guidance represents the view of the National Institute for Health and Clinical Excellence (NICE), which was arrived at after careful consideration of the evidence available. Healthcare professionals are expected to take it fully into account when exercising their clinical judgement. However, the guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer, and informed by the summary of product characteristics of any drugs they are considering.
- Implementation of this guidance is the responsibility of local commissioners and/or providers. Commissioners and providers are reminded that it is their responsibility to implement the guidance, in their local context, in light of their duties to avoid unlawful discrimination and to have regard to promoting equality of opportunity. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties.
- The guideline will assume that prescribers will use a drug's summary of product characteristics (SPC) to inform decisions made with individual patients. This guideline recommends some drugs for indications for which they do not have a UK marketing authorisation at the date of publication, if there is good evidence to support that use. Where recommendations have been made for the use of drugs outside their licensed indications ('off-label use'), these drugs are marked with a footnote in the recommendations.

Implementation of the Guideline

Description of Implementation Strategy

The National Institute for Health and Clinical Excellence (NICE) has developed tools and advice to help organisations implement this guidance. These are available on the [NICE Web site](#) ; see also the "Availability of Companion Documents" field).

Key Priorities for Implementation

The following recommendations have been identified as priorities for implementation.

Principles of Care

- Children and young people with spasticity should have access to a network of care that uses agreed care pathways supported by effective communication and integrated team working.
- If a child or young person receives treatment for spasticity from healthcare professionals outside the network team, this should be planned and undertaken in discussion with the network team to ensure integrated care and effective subsequent management.
- Offer a management programme that is:
 - Developed and implemented in partnership with the child or young person and their parents or carers
 - Individualised
 - Goal focused
- Help children and young people and their parents or carers to be partners in developing and implementing the management programme by offering:
 - Relevant, and age and developmentally appropriate, information and educational materials
 - Regular opportunities for discussion and
 - Advice on their developmental potential and how different treatment options may affect this
- Monitor the child or young person's condition for:
 - The response to treatments
 - Worsening of spasticity
 - Developing secondary consequences of spasticity, for example pain or contractures
 - The need to change their individualised goals

Physical Therapy (Physiotherapy and/or Occupational Therapy)

- All children and young people with spasticity referred to the network team should be promptly assessed by a physiotherapist and, where necessary, an occupational therapist.
- Following treatment with botulinum toxin type A, continuous pump-administered intrathecal baclofen, orthopaedic surgery or selective dorsal rhizotomy, provide an adapted physical therapy programme as an essential component of management.

Intrathecal Baclofen

- Consider treatment with continuous pump-administered intrathecal baclofen¹ in children and young people with spasticity if, despite the use of non-invasive treatments, spasticity or dystonia are causing difficulties with any of the following:
 - Pain or muscle spasms
 - Posture or function
 - Self-care (or ease of care by parents or carers)

Orthopaedic Surgery

- An assessment should be performed by an orthopaedic surgeon within the network team if:
 - Based on clinical findings or radiological monitoring, there is concern that the hip may be displaced.
 - Based on clinical or radiological findings there is concern about spinal deformity

¹ At the time of publication (July 2012), intrathecal baclofen did not have UK marketing authorisation for children younger than 4 years, nor did it have UK marketing authorisation for use in the treatment of dystonia associated with spasticity. Where appropriate, informed consent should be obtained and documented.

Implementation Tools

Audit Criteria/Indicators

Clinical Algorithm

Foreign Language Translations

Patient Resources

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

Staying Healthy

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

National Collaborating Centre for Women's and Children's Health. Spasticity in children and young people with non-progressive brain disorders. Management of spasticity and co-existing motor disorders and their early musculoskeletal complications. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Jul. 54 p. (NICE clinical guideline; no. 145).

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2012 Jul

Guideline Developer(s)

National Guideline Alliance - National Government Agency [Non-U.S.]

Source(s) of Funding

National Institute for Health and Clinical Excellence (NICE)

Guideline Committee

Composition of Group That Authored the Guideline

Group Members: Paul Eunson (*Chair*), Consultant Paediatric Neurologist, Royal Hospital for Sick Children, Edinburgh; Gordon Allan, General Practitioner, Methil, Fife; Liz Barnes, Parent and carer member, Freelance Writer, Trustee of Hemihelp, London; Lucinda Carr, Consultant Paediatric Neurologist, Great Ormond Street Hospital for Children, London; Stephanie Cawker, Clinical Specialist Physiotherapist, Neurodisabilities, Great Ormond Street Hospital for Children, London; Elspeth Dixon, Parent and carer member, Senior Specialist Learning Disability Physiotherapist, Adult Learning Disability Team, Herefordshire; Christina Gericke, Specialist Paediatric Occupational Therapist (retired May 2011), Royal Hampshire Country Hospital, Winchester; Alec Musson, Superintendant Physiotherapist, Paediatric Physiotherapy Team, Leeds Teaching Hospitals NHS Trust; James Robb, Consultant Orthopaedic Surgeon, Royal Hospital for Sick Children, Edinburgh; Trudy Ward, Head of Children's Community Nursing, Sussex Community NHS Trust; Jane Williams, Consultant Community Paediatrician, Nottingham University Hospitals NHS Trust

Financial Disclosures/Conflicts of Interest

All guideline development group (GDG) members' potential and actual conflicts of interest were recorded on declaration forms provided by the National Institute of Health and Clinical Excellence (NICE) (summarised in Appendix B of the full version of the guideline; see the "Availability of Companion Documents"). None of the interests declared by GDG members constituted a material conflict of interest that would influence recommendations developed by the GDG.

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available from the [National Institute for Health and Clinical Excellence \(NICE\) Web site](#) .

Availability of Companion Documents

The following are available:

- Spasticity in children and young people with non-progressive brain disorders: management of spasticity and co-existing motor disorders and their early musculoskeletal complications. Full guideline. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Jul. 298 p. (Clinical guideline; no. 145). Electronic copies: Available in Portable Document Format (PDF) from the [National Institute for Health and Clinical Excellence \(NICE\) Web site](#) .
- Spasticity in children and young people with non-progressive brain disorders: management of spasticity and co-existing motor disorders and their early musculoskeletal complications. Appendices. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Jul. (Clinical guideline; no. 145). Electronic copies: Available in PDF from the [NICE Web site](#) .
- Spasticity in children and young people. NICE pathway. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Jul. Electronic copies: Available from the [NICE Web site](#) .
- Spasticity in children and young people with non-progressive brain disorders. Clinical audit tools. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Jul. (Clinical guideline; no. 145). Electronic copies: Available from the [NICE Web site](#) .
- Spasticity in children and young people with non-progressive brain disorders. Costing report. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Jul. 33 p. (Clinical guideline; no. 145). Electronic copies: Available in PDF from the [NICE Web site](#) .
- Spasticity in children and young people with non-progressive brain disorders. Baseline assessment tool. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Jul. (Clinical guideline; no. 145). Electronic copies: Available in PDF from the [NICE Web site](#) .

- Spasticity in children and young people with non-progressive brain disorders. Implementation advice. London (UK): National Institute for Health and Clinical Excellence (NICE); 2012 Sep. 40 p. (Clinical guideline; no. 145). Electronic copies: Available in PDF from the [NICE Web site](#) .
- The guidelines manual 2009. London (UK): National Institute for Health and Clinical Excellence (NICE); 2009 Jan. Electronic copies: Available in PDF from the [NICE Archive Web site](#) .

Patient Resources

The following is available:

- Spasticity in children and young people. Information for the public. 2012 Jul. Electronic copies: Available from the [National Institute for Health and Clinical Excellence \(NICE\) Web site](#) . Also available in Welsh from the [NICE Web site](#) .

Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

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This NGC summary was completed by ECRI Institute on January 21, 2013. This summary was updated by ECRI Institute on October 21, 2016 following the U.S. Food and Drug Administration advisory on opioid pain and cough medicines combined with benzodiazepines. This summary was updated by ECRI Institute on February 15, 2017 following the U.S. Food and Drug Administration advisory on general anesthetic and sedation drugs.

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